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Case Report

CASE REPORT

Bilateral Orbital Pseudotumor in a 3-Year-Old Child: A Case Report

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ABSTRACT

Orbital pseudotumor is a rare, idiopathic, inflammatory process within the orbit. Early diagnosis is essential to prevent corneal, retinal and extraocular muscle damage. This article describes the presentation and clinical course of a 3-year-old male with bilateral orbital pseudotumor.

KEYWORDS: orbital pseudotumor, children, Ethiopia

INTRODUCTION

Orbital pseudotumor, also known as Idiopathic Orbital Inflammatory Syndrome, is a benign, non-infectious, non-specific inflammatory condition of the orbit in which no local or systemic cause can be identified [1]. The disease is rarely observed in children. In this case report, we describe the management of a 3-year-old boy with orbital pseudotumor at University of Gondar Hospital, Ethiopia. Informed consent was obtained from his family before publication of this report.

CASE REPORT

A 3-year-old Ethiopian boy was referred to Gondar University hospital with a 1 year history of bilateral eye protrusion. The patient's family first observed a painless swelling of both eye lids. The patient developed watery eye discharge and redness of the eye over 2 months but denied itchiness or pain. He endorsed photophobia but no change in vision. He had no double vision or limitation of eye movements. He had no fever, loss of appetite or decreased activity. He had no history of trauma or surgery. He had no headache, vomiting or weight loss. His vital signs, anthropometry and systemic examination were normal. He had bilateral proptosis, mild bilateral ptosis, conjunctival injection and mild chemosis (Fig. 1). His pupils were isocoric, round and reactive to light. Extraocular muscle movement was −3 restricted in all eye direction. Funduscopic examination after pupillary dilation showed +1 disc edema. His complete blood count, sedimentation rate, urinalysis, serum electrolyte and liver and renal function tests were normal. His

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Fig. 1. Eyelid swelling and proptosis.

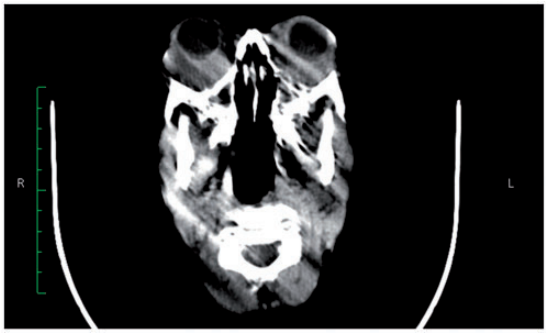

Fig. 2. Orbital CT scan, axial view, showing a bilateral, symmetrical proptosis and a bilateral symmetrical hypertrophy of extraocular muscles and tendons.

HIV antibody test was non-reactive. His peripheral morphology showed normocytic normochromic red blood cells and no peripheral blast cells. His chest radiograph and abdominal ultrasound were normal. His skull radiograph showed diffuse, bilateral enlargement of the periorbital tissue. Computed tomography (CT) scan of the orbit showed bilateral proptosis with bilateral symmetric hypertrophy of all extraocular muscles and tendons but no adjacent bone destruction (Fig. 2). Under general anesthesia, biopsy was taken from his right inferior inflammatory mass.

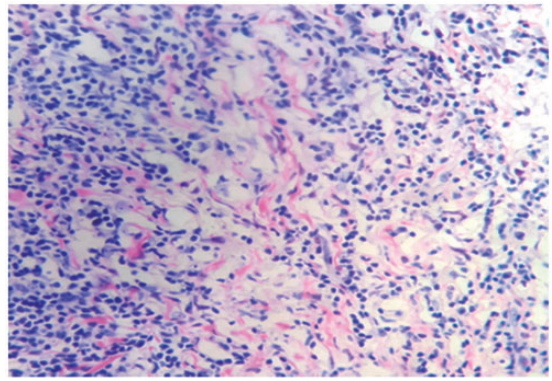

Fig. 3. High power (40×) microscopy showing dense lymphoplasmacytic infiltrates with areas of collagen bundles.

Fig. 4. After one month of prednisolone therapy, the patient responded with decreased proptosis, eye lid swelling and conjunctival injection.

Grossly one irregular gray white tissue fragment was received by the pathology department. The received tissue was processed by automatic processor and stained by standard Hematoxylin and Eosin stain. Light microscopy showed numerous chronic inflammatory cells, mainly lymphocytes, plasma cells and few histiocytes with adjacent fibroblasts depositing collagen bundles. No necrosis, granuloma or malignant cells were seen (Fig. 3).

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The child was admitted to the pediatric ward with an initial diagnosis of bilateral orbital mass and acute bacterial conjunctivitis. Tetracycline eye ointment was administered three times daily for 5 days with no effect. Oral prednisolone was started at 1 mg/kg daily on the second day just after the biopsy results were reported. A reduction of proptosis, lid edema and conjunctival injection were observed within 48 h of treatment (Fig. 4). The child was discharged from the pediatric ward after 10 days of treatment. He was appointed to the pediatric follow-up clinic, where the prednisolone was tapered after 6 weeks of treatment because of complete resolution of symptoms.

DISCUSSION

Orbital pseudotumor was first described by Birch-Hirschfield in 1905. Although it is widely considered to be idiopathic, unrecognized microorganisms, minor trauma and chronic irritation have been suggested as likely triggers [2, 4]. Orbital pseudotumor is the third most common ophthalmologic disease of the orbit, accounting for 8–11% of all orbital tumors. It can affect both sexes and any age or ethnicity but is rare in childhood [3, 5, 6].

Children present with sudden or slow and progressive onset of pain, eyelid edema, proptosis and subconjunctival hemorrhage. Vision loss, photophobia, diplopia, ptosis and motility restriction are common. Headache, vomiting and weight loss are more common in children than in adults. It is also more common in children to present with bilateral involvement, iritis and a history of trauma preceding the inflammation [3, 6, 7].

The cause of orbital pseudotumor is unknown. Several theories suggest an immune-mediated process that compresses the orbital structures and results in a mass effect. Infectious, genetic and environmental factors also have been suggested [8].

Orbital pseudotumor can be confused with acute orbital cellulitis, thyroid ophthalmopathy, orbital vasculitis, Wegener granulomatosis, sarcoidosis and malignant tumors, such as lymphoma and rhabdomyosarcomas [6, 9]. Laboratory tests in orbital pseudotumor are often normal, but may reveal peripheral eosinophilia, elevated sedimentation rate and elevated serum antinuclear antibody test. Orbital CT, magnetic resonance imaging and B-scan ultrasonography can be useful in excluding other etiologies [2]. Histologic findings can vary, but a mixture of plasma cells, macrophages, eosinophils, mature lymphocytes and polymorphonuclear cells are typically reported. Occasionally, the density of fibrovascular stroma is increased [10].

Oral systemic corticosteroids are the mainstay treatment. Rapid response to steroids also is considered diagnostic. Prednisone 1–1.5 mg/kg per day in children leads to resolution of pain and proptosis within 24–48 h. When the amount of collagenous connective tissue increases as seen in more chronic forms of orbital pseudotumor, the sensitivity to corticosteroids decreases. Low-dose radiotherapy can be indicated when corticosteroids fail or are medically contraindicated, or for recurrences when the patient is already receiving corticosteroid therapy [2, 8, 11].

Orbital pseudotumor is a rare inflammatory condition, occasionally presenting chronically with bilateral involvement in children. It is often misdiagnosed as orbital cellulitis or orbital mass with conjunctivitis. Because this disorder affects vision, it is crucial to diagnose and treat promptly.

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